

A guide to treatment with Ravicti[®] ▼



This brochure is only for patients who have been prescribed Ravicti (Glycerol phenylbutyrate) when a healthcare professional wishes to provide this as an information resource

▼ This medicine is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may get. See page 5 for how to report side effects.

THIS IS RAVICTI

Ravicti® (glycerol phenylbutyrate) is a treatment used to treat patients of all ages with urea cycle disorders (UCDs). The medicine is used to reduce the amount of ammonia in the blood in order to reduce the risk of neurological consequences.

Ravicti is an oral liquid, which means it's taken by mouth. It is usually taken 3 times a day by oral syringe in equal doses. It must be used with dietary protein restriction and, in some cases, dietary supplements. Because every patient's needs are different your healthcare team will come up with a personalised plan on how to best manage your ammonia levels.

Each unit of Ravicti contains a glass bottle and a bottle adapter. It does not require any special storage conditions.

After the first opening of the bottle Ravicti must be used within 14 days and the bottle and its contents discarded, even if not empty.

This brochure does not replace the advice you'll get from your healthcare team. If you have any questions about your disease and treatment, please talk to your doctor.

HANDLING YOUR UREA CYCLE DISORDER

Apart from medical treatment, individualised dietary treatment is one of the cornerstones to help control high levels of ammonia associated with a UCD.

Low-protein diet

It's important to limit protein when living with a UCD. However, limiting it too much may harm your body at the same time, because we need it to promote growth and development. Common triggers, like being stressed or getting sick, can increase ammonia to toxic levels despite your best efforts to balance protein intake. Low-protein doesn't automatically mean "boring" food. There are plenty of tasty dishes low in protein. You can also modify your favourites to lower their protein level.

Amino acid supplements

Essential nutrients are needed to grow and maintain your health. Amino acid supplements fill the gap when the amount of protein you can eat is restricted and supply you with essential vitamins and minerals.

① A dietitian who is part of your healthcare team can help decide which supplements may be right for you. The dietitian can also help you with dietary advice such as low protein recipes, cooking etc.

YOUR TREATMENT WITH RAVICTI



Remember to read the patient information leaflet thoroughly before taking Ravicti.

How can Ravicti help treat a urea cycle disorder?

One of the most important goals of managing a urea cycle disorder (UCD) is to lower the amount of ammonia in your blood to a safe and normal level. Ammonia levels can go up and down during the day and can be harmful to the body when they are too high. Ravicti may help you by controlling elevated levels of ammonia. Ravicti is an oral medicine, which means it's taken by mouth. It has little or close to no taste. It can be used to treat patients of all ages for chronic management of elevated levels of ammonia caused by a UCD.

Who should not take Ravicti

Do not take Ravicti if you are allergic to phenylbutyrate. Call your doctor or go to the nearest hospital if you have symptoms of an allergic reaction, such as wheezing, shortness of breath, cough, low blood pressure, flushing, nausea or rash while taking Ravicti.

What are the possible side effects of Ravicti?

Every medical treatment may cause side effects, but which side effects that occur varies from person to person. The most common side effects with Ravicti include: diarrhoea, flatulence, headache, nausea, vomiting, tiredness, decreased appetite, skin smells abnormal and dizziness.

In children less than 2 months of age

Reflux of stomach contents, diarrhoea, rash, reduced number of red blood cells, increased number of platelets, flatulence, constipation, increased liver enzyme and poor feeding.

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in this leaflet. Adverse events should be reported. Reporting forms and information in the UK can be found at www.mhra.gov.uk/yellowcard and in Ireland they can be found at www.hpra.ie. Adverse events should also be reported to Immedica Pharma UK Ltd by email: safety@immedica.com

When to get medical help right away

The breakdown of Ravicti produces the byproduct phenylacetate, which may cause nervous system side effects. Call your doctor or get medical help right away if you have any of these symptoms while taking Ravicti:

- sleepiness
- lightheadedness
- change in taste
- problems with hearing
- confusion
- problems with memory
- worsening of numbness
- tingling, or burning in your hands or feet
- headache, tiredness, nausea or vomiting

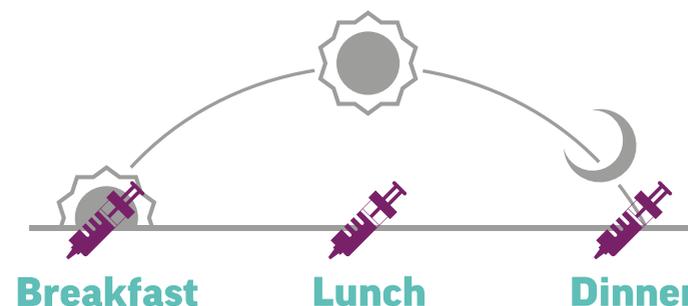
Before you take Ravicti

Tell your doctor:

- if you have liver or kidney problems, pancreas or bowel (intestine) problems, or any other medical conditions
- if you are pregnant or plan to become pregnant. It is not known if Ravicti will harm your unborn baby
- if you are breastfeeding or plan to breastfeed. It is not known if Ravicti passes into your breast milk. Breastfeeding is not recommended during treatment with Ravicti. Your doctor can council you about the best way to feed your baby if you take Ravicti
- if you are taking, have recently taken or might take any other medicines, including contraceptives

HOW TO TAKE RAVICTI

Your doctor will tell you how much Ravicti you should take each day. Your daily dose will depend on your size and weight, the amount of protein in your diet, and your overall urea cycle disorder condition.



Ravicti is usually taken 3 times a day in equal doses, with food or formula. Take or give Ravicti exactly as your doctor has prescribed it which can be more than 3 times each day. In small children, this may be 4 to 6 times a day. There must be at least 3 hours between each dose. Your doctor may change your dose if needed. If too much Ravicti is taken, call a doctor and go to the nearest hospital emergency room right away.

Missing a dose

For adults: If you forget a dose, take the missed dose as soon as you remember. If the next dose is in less than 2 hours, then skip it and take your next dose as normal.

For children: if the next dose is in less than 30 minutes, then skip it and give the next dose as normal.

Do not take a double dose to make up for a forgotten dose. If you have further questions on the use of this medicine, ask your doctor or pharmacist.

On the next page, you will find directions for taking Ravicti.



Open the bottle of Ravicti. To open, push down on the cap while twisting it to the left.



Twist the blue AdaptaCap Bottle Adapter onto the bottle. You do this by turning it to the right. Make sure it is tightly on the bottle.



Place the tip of the oral dosing syringe into the AdaptaCap Bottle Adapter. Your doctor or pharmacist will provide you with the correct syringes.



Turn the bottle upside down with the oral dosing syringe still inserted.



Get the medicine into the oral dosing syringe by pulling the plunger back. Pull until the first line of the black rubber stopper is even with the dosing mark. The dosing marks are on the barrel of the oral dosing syringe. Match the dosing mark to the amount of medicine prescribed by your doctor.

EXAMPLE: If your prescribed dose is 3 ml, the first line of the rubber stopper should be even with the 3 ml dosing mark on the barrel of the oral dosing syringe.

NOTE: Pull the plunger slowly. This will help keep large air bubbles from forming in the medicine.



Turn the bottle back to its upright position.



If there are large air bubbles, push the medicine back into the bottle. Do this slowly. Draw up the medicine again (repeat steps 3-6).



Place the oral dosing syringe into the mouth. Push the plunger to squirt the medicine into the mouth. Make sure all of the medicine is pushed out of the oral dosing syringe. Put the cap on the oral dosing syringe.



After taking Ravicti, it is suggested to drink liquid or eat some food to help make sure all of the medicine is swallowed.



Close the tab on the AdaptaCap Bottle Adapter. Make sure it is closed tightly.

IMPORTANT TO REMEMBER:

- Take the exact amount of Ravicti as instructed by your doctor.
- Take Ravicti with meals, in equally divided doses.
- When dosing Ravicti be sure to only use the oral syringes provide by your pharmacist.

NOTE: You should use 1 oral syringe for all doses each day, but after the last dose of each day, dispose of the oral syringe.

- Do not rinse the AdaptaCap Bottle Adapter or oral dosing syringe between doses.

If you take too much Ravicti, call your doctor or go to the nearest hospital emergency room right away.

Do not stop taking Ravicti unless your doctor tells you to stop.

- If you miss a dose of Ravicti, take it as soon as possible.
- Do not take 2 doses of Ravicti at the same time.
- If you miss more than 1 dose and are not feeling well, call your doctor or nurse right away.

If you still have questions about how to take Ravicti with an oral dosing syringe, please contact your doctor for more information.



Immedica Pharma UK Ltd
The Townhouse
114-116 Fore Street
Hertford, Herts, SG14 1AJ